Thoracoscopic treatment of a rare bilateral extralobar lung sequestration in a 3-years old girl

Cosimo Bleve,[1] Maria Luisa Conighi,[1] Diego Biondini,[2] Pier Luca Ceccarelli,[2] Leonardo Giarraputo,[3] Sergio Savastano,[3] Salvatore Fabio Chiarenza[1]

[1]Department of Pediatric Surgery and Pediatric Minimally Invasive Surgery and New Technologies, San Bortolo Hospital, Vicenza; [2]Department of Pediatric Surgery, University of Modena and Reggio Emilia, Policlinico of Modena; [3]Department of Radiology, San Bortolo Hospital, Vicenza, Italy

Abstract

Majority of sequestrations fall into two categories: Intra-Lobar (ILS) and Extra-Lobar (ELS). Rarely the abnormal lung could be attached to the gastrointestinal tract, Bronchopulmonary Foregut Malformation (BPFM). We described a case of a girl of 3-years-old with antenatal diagnosis of left intrathoracic mass of the inferior lobe. Postnatal Computed-Tomography (CT) revealed a bilateral ELS with an isthmic bridge crossing the vertebral spine. She follows a MRI follow-up at 18months/30months confirming the lesion. Before surgery, a three-dimensional-CT-angiography was performed to study the mass, its blood supply and to plan surgery. She underwent to thoracoscopic resection. Two aberrant blood vessels were dissected from the thoracic aorta and ligated. The postoperative course was uneventful. She was discharged after 3 days. The rarity of our case is due to the bilateral extension. An appropriate preoperatory imaging study is necessary for the success of surgery while thoracoscopy is particularly appropriate in surgical treatment.

Introduction

Lung sequestration is one of the rare thoracic congenital anomalies. It is characterized by an area of dysplastic and non-functional lung tissue that receives its blood supply from aberrant systemic arteries and is not in normal continuity with the tracheo-bronchial tree.[1] It is usually classified as either Intra-Lobar Sequestration (ILS), which is located within a lung lobe and shares the visceral pleura with the corresponding lung lobe, or Extra-Lobar Sequestration (ELS), which is a separate mass of lung parenchyma enclosed entirely by a separate pleural envelope.[2] ILS accounts for 75% of pulmonary sequestration, while ELS constitutes the remaining 25%. Although ILS almost always occurs within the lower lobe (98%) and more often in the left lung (55%), ELS is typically found in the posterior costodiaphragmatic sulcus.[1-3] ILS is four times more common than ELS. ILS has an equal incidence in both males and females whereas ELS is present in males in >80% cases.[4] Extralobar-pulmonary sequestration is a rare congenital lung anomaly which represents the second most common lung malformation in children, accounting for 0.1 to 1.8% of cases.[5,6] Its origin remains uncertain; however, the available evidence suggests that it results from accessory lung tissue arising from the foregut. ELS is located in the mediastinum in approximately 5% of cases.[7] Our case presented ELS with extension from the left to the right hemithorax, with horseshoe shape. It was successfully treated by monolateral left thoracoscopy. By a revision of the literature, this is the only case reported in pediatric population of a median intra-thoracic extra-lobar sequestration.

Case Report

A 3-years-old girl was diagnosed with fetal ultrasound and confirmed by magnetic resonance imaging as a mediastinal mass lesion. She was delivered by cesarean section at 33 weeks' gestation with a birth weight of 3,300g. The perinatality was regular.
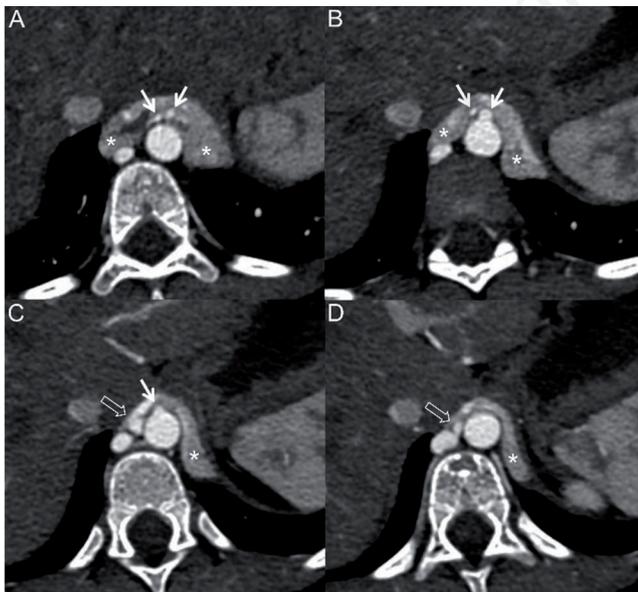
Postnatal CT at 3th month of life showed an ELS; the follow-up was performed with two MRI at 18 and 30 months of life. Health condition of the girl were good. No episodes of lower respiratory tract infection were recorded.

Preoperatory imaging study

The three-dimensional CT-angiography at 2-years of age showed the ELS located at the thoraco-abdominal passage, in the posterior mediastinum, in the distal sectors, with a "saddlebag" appearance where the two components have a diameter of 25 mm (each of 25 mm, totally 50 mm) with isthmic connection. This structure is included between the anterior esophagus and the thoracic aorta in the posterior sectors between the diaphragmatic pillars. In the contrast phase it presented a rich enhancement whose arterial supply appears to be of a systemic type ensured by two branches originating from the thoracic aorta; the cranial artery measured 3.5 mm in diameter whereas the caudal artery 2.5 mm (Figure 1A-1D). Both the systemic arteries originated from the ventral aspect of the thoracic aorta just above the diaphragmatic hiatus (Figure 2A-2B). Venous drainage is ensured through a single branch afferent to the azygos vein. The right side of the lesion contains a tiny hypoattenuating tubular structure, likely consistent with a rudimental bronchus (Figure 1A).

Surgical treatment

At 3-years old, the girl underwent to thoracoscopic resection and asportation of lung sequestration. The patient was on right quite prone decubitus with the left arm blocked up over the head. A 5 mm 30° optic was inserted at the lower corner of the left scapula. We started with CO2 insufflation into the hemithorax to partially collapse the lung, maintaining a pressure of 6-7 mmHg. Other two 5 mm trocars were positioned along the same line as the previous one, using then the median access for the optic and then switch to 3D vision. The posterior portion of the hemithorax is explored: diaphragm and costophrenic sinus were regular; were
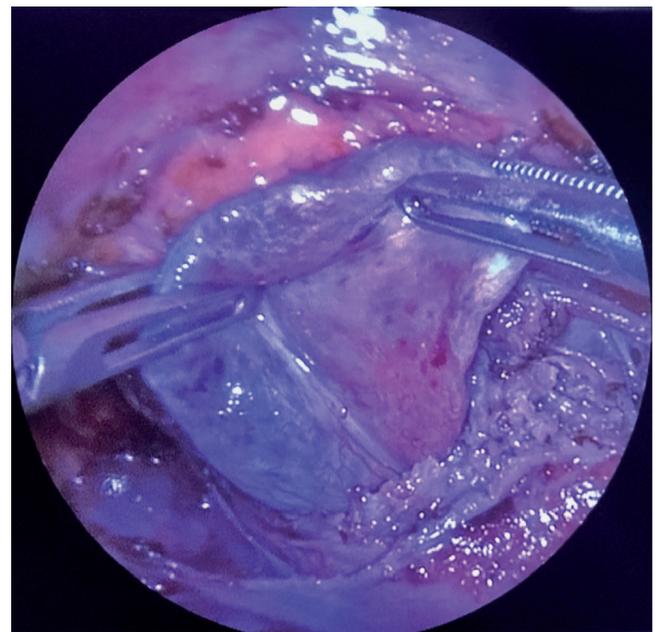
visualized the column, aortic arch and thoracic aorta; the mediastinal parietal pleura, anterior to the aorta, passing through the diaphragm has a hyperemic and thickened area. A possible underlying lesion appears to be appreciated in this area, then the mediastinal parietal pleura is incised anteriorly to the thoracic aorta. Carefully was start the dissection of a cystic lesion at first appearance, with soft consistency that starts on the left side of the posterior mediastinum and extends medially between the distal tracts of the thoracic aorta posteriorly and the esophagus anteriorly. To facilitate the operator's maneuvers, a third trocar 5 mm is positioned. The lesion was close to the esophagus. Therefore, intraoperative endoscopy was performed to allow the exact identification of the esophagus, the relationships with the lung sequestration, and to exclude that it may be an esophageal duplication. The two anomalous systemic arteries originating from thoracic aorta were dissected, ligated and divided. The dissection of the lesion was conducted to the median portion of the mass, which was sectioned
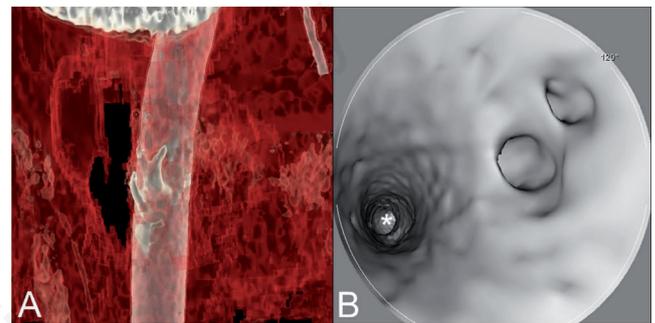
Figure 2. A) The two systemic arteries arising from the anterior wall of the aorta are well appreciable on volume rendering of the descending aorta. B) Intravascular virtual navigation of the aorta (inferior view). Ostia of the systemic arteries are nicely appreciable on the right side (*aortic isthmus).

Figure 1. A crescent pulmonary sequestration (*) circumscribes the ventral aspect of the lower third of the descending aorta. The pulmonary sequestration is supplied by two systemic arteries, distinctly arising from the anterior wall of the thoracic aorta; The cranial artery measured 3.5 mm in diameter whereas the caudal artery 2.5 mm (black arrow). The lesion drains into the azygos vein through a large vein (white arrow).

Figure 3. ELS: isolated accessory lung parenchyma dissected from its separate pleural envelope.

with Ligasure (Figure 3). The small remnant on the right side appeared ischemic and so was left in place. A chest tube was positioned for 24 hours. The pathological diagnosis was ELS. The patient's postoperative course was uneventful without symptoms after 1-year follow-up.

Discussion

The antenatal discovery of a cystic or solid lung malformation opens a range of diagnosis, with the most frequent being congenital pulmonary airway malformation and Bronchopulmonary Sequestration (BPS). The distinction between these two entities is not so clear with the discovery of an increasing number of "hybrid" lesions as reported by Davenport *et al*.[8] The natural fetal evolution of these lesions may either tend toward enlargement, or to steady state, or even to partial/complete regression.[9] Surgery presents an increase in risk of operative complications in symptomatic patients; so elective surgery is aimed to be performed before appearance of symptoms, such as respiratory distress or infection. The average time of symptoms onset is reported to be at the age of about 10 months, considering both infants diagnosed prenatally and those who were not.[10] Thus, most pediatric surgeons generally plan elective surgery in infants with congenital lung lesions at the age of 6–12 months.[11] Pulmonary sequestration constitutes approximately 0.15–6.4% of all congenital pulmonary malformations. Nowadays, according to the most accepted theories, ILS and ELS presented a different pathogenesis. ILS is considered an acquired disease associated with bronchial obstruction, pneumonia and pleuritis, with the development of a systemic arterial supply secondary to angiogenic growth factors that are activated by recurrent infections. It is also considered a variant of Congenital Cystic Adenomatoid Malformations (CCAM). ELS is considered a real congenital disease. Embryologically in utero, an accessory lung bud develops from the ventral aspect of the primitive foregut. This bud, a pluripotential tissue, migrates in a caudal direction with the normally developing lung. This lung bud receives its blood supply from aberrant arteries arising from the aorta and cover the primitive foregut. Accessory lung bud ultimately develops into sequestered tissue that is not attached to the pulmonary arterial blood supply, no connected to the normal bronchial airway architecture. This sequestrated tissue may ultimately become cause of life threatening hemorrhage and recurrent infections.[2] The vascular supply for both ILS and ELS arises usually, directly from or the lower thoracic or upper abdominal aorta. The majority of sequestration have a single aberrant vessel, although up to a third present multiple vessels as in our case. The venous drainage is usually in the left atrium but an abnormal drainage to the right atrium, cava vein and azygous vein (as in our patient) is reported. In our patient, the blood supply to the sequestered portion was from two arteries arising from descending aorta. ELS could be associated more than ILS to other anomalies as CDH, vertebral anomalies, congenital heart disease, lung hypoplasia, colonic duplication. In our case ELS was isolated. Conventional CT does not always demonstrate the systemic artery, especially if the vessels are less then 1mm. Preoperative three-dimensional CT-angiography is useful for two principle objectives for the investigation of a suspected case of pulmonary sequestration: i) to confirm the presence of an anomalous systemic arterial supply to the sequestered lung and ii) to distinguish pulmonary sequestration from other lung opacities.[2] The ability to rotate 3D VR images and display them in any orientation is helpful in discerning the relationships that exist within aberrant angioarchitecture.[12-14] In our case the 3D CT-angiography allowed

to plan the surgical approach. Surgical resection is the conventional standard treatment for Pulmonary Sequestration (PS) to prevent possible infection, congestive heart failure, and hemoptysis. The treatment is still controversial. Is considered accepted that PS causing symptoms should be resected. About ELS conservative treatment is one possibility for asymptomatic patients considering that these lesions rarely produce symptoms. What must be considered is that a number of lesions diagnosed as sequestration are really "ibrid" lesion, with CCAM-type morphology. PS manifested as an underlying cancer predisposition syndrome and an innate propensity to undergo malignant degeneration. Lu *et al*.[15] reported in their paper a case of Atypical Adenomatous Hyperplasia (AAH) in ELS. AAH is considered a pre-cancerous lesion; it is involved in the early stage of a complex multistep carcinogenesis of pulmonary adenocarcinoma. They reported also by a revisions of the literature, less than ten cases of carcinoma arising in PS, including four cases of pulmonary adenocarcinoma, also if only one arising in ELS. Belchis *et al*.[16] hypothesized that the development of carcinoma in the sequestrations may be multifactorial and may include chronic inflammation and irritation. For this reason we consider resection in all cases the most appropriate therapy mostly nowadays when minimally invasive technique are emerging in the treatment of this pathology. What is essential for surgery is to identify and control aberrant angioarchitecture. Careful preoperative identification of systemic arterial supply and venous drainage from a pulmonary sequestration is necessary to avoid hemorrhage during surgery. In our case the thoracoscopic approach on the left side was the correct choice because we can identify and control the two arteries suppling the ES. We removed about 75% of the mass leaving the right portion with is venous drainage in the azygos vein which appeared ischemic. The rationale of this choice is the same of embolization of the systemic feeding vessel, reported as surgical alternative to surgical resection with regression of the tissue.[17,18]

Conclusions

In conclusion, 3-dimensional thoracoscopic surgery is useful for patients with ELS. Resection of the ELS through the left side was a reasonable approach for this patient because the ELS was located at the lower posterior mediastinum and the aberrant vessels were near the hiatus on the descending aorta. The laparoscopic approach could be considered for the site of ELS but probably this choice should not be suitable because ELS was large with the aberrant vessels far from the hiatus. We suggest that this approach might be adapted in other similar cases of ELS.

References

1. Mendeloff EN. Sequestrations, Congenital cystic adenomatoid malformations, and congenital lobar emphysema. Semin Thorac Cardiovasc Surg 2004;16:209-14.
2. Long Q, Zha Y, Yang Z. Evaluation of pulmonary sequestration with multidetector computed tomography angiography in a select cohort of patients: A retrospective study. Clinics 2016;71:392-8.
3. Savic B, Birtel FJ, Tholen W, et al. Lung sequestration: report of seven cases and review of 540 published cases. Thorax 1979;34:96–101.
4. Hussain S, Ahmed S, Hassan Shah SAH, Tarar SH. A rare cause of recurrent chest infection in children-bronchopul-

OPEN ACCESS

monary sequestration. J Pak Med Assoc 2017:67:311-3.

5. Chouikh T, Berteloot L, Revillon Y, et al. Extralobar pulmonary sequestration with combined gastric and intradiaphragmatic locations. Pediatr Pulmonol 2014;49:512–4.

6. Bush A. Prenatal presentation and postnatal management of congenital thoracic malformations. Early Hum Dev 2009;85:679–84.

7. Miyagi H, Honda S, Hamada H, et al. One-Stage Laparoscopic Surgery for Pulmonary Sequestration and Hiatal Hernia in a 2-Year-Old Girl. Eur J Pediatr Surg Rep 2018;6:e11–4.

8. Davenport M, Warne SA, Cacciaguerra S, et al. Current outcome of antenatally diagnosed cystic lung disease. J Pediatr Surg 2004;39:549–56.

9. Divjak N, Vasseur Maurer S, Giannoni E, et al. Bronchopulmonary Sequestration with Morbid Neonatal Pleural Effusion despite Successful Antenatal Treatment. Front Pediatr 2017;5:259.

10. Stanton M, Njere I, Ade-Ajayi N, et al. Systematic review and meta-analysis of the postnatal management of congenital cystic lung lesions. J Pediatr Surg 2009;44:1027–33.

11. David M, Lamas-Pinheiro R, Henriques-Coelho T, Prenatal and postnatal management of congenital pulmonary airway malformation. Neonatology 2016;110:101–15.

12. Choe J, Goo HW. Extralobar pulmonary sequestration with hemorrhagic infarction in a child: preoperative imaging diagnosis and pathological correlation. Korean J Radiol. 2015 May-Jun;16(3):662-7.

13. Fumino S, Iwai N, Kimura O, et al. Preoperative evaluation of the aberrant artery in intralobar pulmonary sequestration using multidetector computed tomography angiography. J Pediatr Surg 2007;42:1776–9.

14. Johnson PL, Fishman EK, Duckwall JR, et al. Interactive three-dimensional volume rendering of spiral CT data: current applications in the thorax. Radiographics 1998;18:165–87.

15. Lu L, Zhang C, Cheng Y. Solitary AAH Arising from Extralobar Sequestration in A Less Than 3-year-old Boy: A Case Report. Zhongguo Fei Ai Za Zhi 2017;20:787-8.

16. Belchis D, Cowan M, Mortman K, et al. Adenocarcinoma arising in an extralobar sequestration: a case report and review of the literature. Lung Cancer 2014;84:92–5.

17. Tokel K, Buyvant F, Varan B. Coil embolization of pulmonary sequestration in two infants: a safe alternative to surgery. Am J Roentgenol 2000;175:993-6.

18. Lee KH, Sung KB, Yoon HK, et al. Trans-catheter arterial embolization of pulmonary sequestration in neonates: long-term follow-up results. J Vasc Intervent Radiol 2003;14:363-7.